

Posterior urethral valves

Originally described by Hugh Hampton Young in 1919

Males only

Incidence 1:5000

Possibly higher incidence in Down's syndrome; No a/w VACTERL

Young originally described 3 types, I II and III

- | | |
|--------|---|
| Type 1 | 95% of valves
Bicuspid valves from veru, through membranous urethra (zone of EUS) to anterior urethral wall |
| Type 2 | Now known not to be a/w posterior valves
Hypertrophied band of tissue in posterior urethra a/w dysfunctional voiding |
| Type 3 | 5% of valves
Sheet membrane with central aperture. Probably a variation of type 1 |

Now believed that all patients have a type 3 iris-shaped deformity, termed a congenitally obstructing posterior urethral membrane (COPUM), which is converted to a 'type 1' deformity by the passage of a catheter after birth. Typically arise at 7 weeks gestation due to abnormal interaction of mesonephric duct confluence and urogenital sinus. Early severe obstruction causes bladder outflow obstruction (valve bladder), upper tract dilatation and renal dysplasia. Later mild obstruction a/w bladder outflow obstruction without dysplasia.

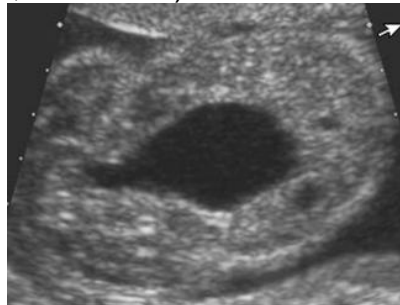
Presentation

Prenatally

Prenatal diagnosis in 80% (approximately 50% detected at routine second trimester scan; remainder later for obstetric indications)

Diagnosed on USS

Distended bladder and dilated upper tracts (DD = urethral atresia (fatal), prune belly and severe bilateral VUR)
Occasionally keyhole sign (dilated bladder and proximal urethra; see below)



Oligohydramnios, pulmonary hypoplasia, moulding defects (Potter's facies, talipes etc.)

Potter's facies (4)

Flattened nose

Recessed chin (hypognathism)

Prominent epicanthal folds

Low-set ears

Predictors of poor functional outcome and early renal failure:

Detection before 24 weeks
Thickened bladder wall
Echobright kidneys (dysplastic)
Oligohydramnios

Postnatally

AUR, palpable abdominal mass, listlessness in neonates

UTI, growth retardation in older children

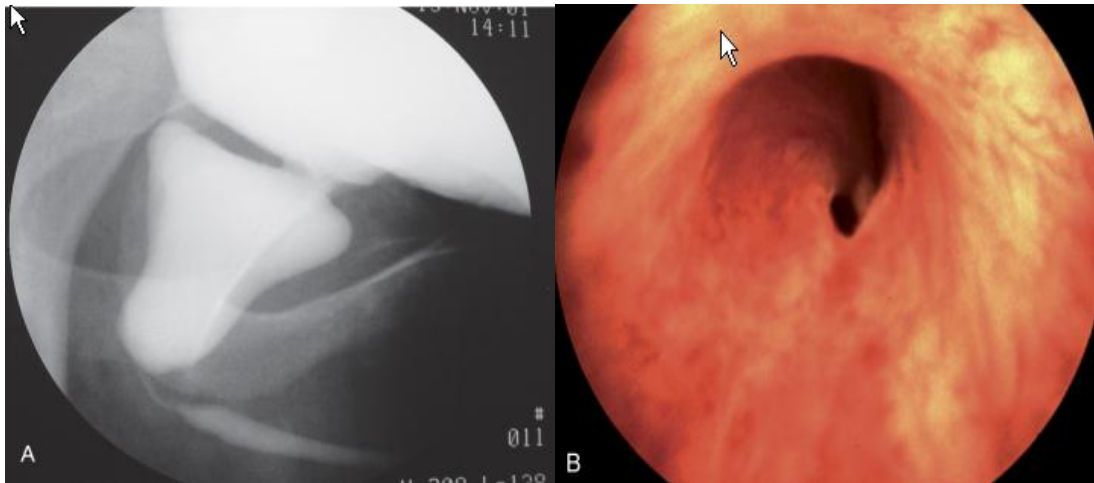
Diagnosis

USS Dilated upper tracts, thick-walled bladder,
occasionally dilatation of proximal urethra

MCU Diagnostic

Dilated proximal urethra with 'sail in the wind'
appearance of valve; dilated trabeculated bladder;
VUR in ~ 50% cases

Perineal USS – research tool only



Management

Prenatally

Fetal vesico-amniotic shunting

Theoretical benefit from animal studies in preventing
decline in renal function and correcting oligohydramnios
RCT never been performed and long-term follow-up
difficult, therefore ? beneficial in humans

Results not encouraging – mortality 40-60% (Fetal
Surgery Register, Elder 1987) but performed on patients
with very poor prognosis

May have limited role

Elective pre-term delivery

Theoretical benefit – earlier treatment in late onset cases
but no evidence base

Late termination of pregnancy

Often for oligohydramnios with marked dilatation

Postnatally

Correction of electrolyte abnormalities *first* followed by early
incision of valves

Correction of electrolyte abnormality

Urethral or suprapubic catheter drainage 2-7 days
5-8F infant feeding tube (Foley balloon can block ureteric orifices)

Vesicostomy for severe renal impairment or when neonatal endoscopic instruments unavailable

Endoscopic valve ablation

4 and 8 o'clock positions

With increasing diameter of urethra:

- a. Whittaker hook
- b. Neonatal cystoscope and bugbee electrode
- c. Paediatric resectoscope and loop

Prognosis

Poor prognostic features in COPUM

Antenatally

Oligohydramnios
Early pre-natal detection < 24 weeks
Thickened bladder wall
Echobright kidneys
High B2 microglobulin in urine

Postnatally

Early presentation < 1 month
Bilateral VUR
Impaired renal function (see below)
Proteinuria
Daytime incontinence (after 5 years old)
UDS showing poor compliance or detrusor failure

Renal failure

30% impaired renal function [primary dysplasia, BOO, VUR and UTI and persistent bladder dysfunction 2' to valve bladder]

Predicting renal failure on follow-up:

Oligohydramnios
Early prenatal detection
Clinical presentation within 6 months
Proteinuria
Bilateral VUR**
Impaired continence > 5 yrs

** recognised good prognosis features ('pop-off' mechanisms) include unilateral VUR with dysplasia (aka VURD: massive reflux into non-functioning kidney preserves function in other kidney), ascites, perinephric urinoma and bladder diverticulum

Valve bladder

Features

Poor sensation
Hypercontractility
Poor compliance
Bladder neck hypertrophy
Concentrating defect leading to high urine output (which increases cycling - therefore more upper tract)

dysfunction. To avoid overnight cycling Koff popularised concept of overnight catheterisation)
Typically high resting intravesical pressures, poor emptying and incontinence, leading to eventual bladder atonicity
UDS findings
10% detrusor overactivity (young)
65% poorly compliant high pressure bladder
25% myogenic failure (old)
Managed by double void and ISC, but compliance poor due to preserved sensation. May require augmentation + mitrofanoff

Other urethral abnormalities

Anterior urethral valves

Incidence 1:40,000 (8x less common than posterior valves)
40% bulbar; 30% penoscrotal; 30% penile
No upper tract pathology
Diagnosis with MCUG
Mx = endoscopic incision

Urethral diverticulum

Usually wide-mouthed at penoscrotal junction
Distal lip may give obstruction
Usually UTI or obstruction
Conservative or excision/grafting

Megalourethra

Dilatation without obstruction
a/w absence of corpus spongiosum
may be part of prune belly syndrome

Cowper's gland cysts

Arise level with urogenital diaphragm (EUS)
Dilatation can cause urethral obstruction

Urethral duplication (Shotgun urethra)

Sagittal (over and under) common
Typically orthotopic urethra with accessory dorsal – usually excised
Collateral (side by side) uncommon

Posterior urethral polyps

Fibroepithelial and pedunculated – excise

Cobbs collar

Controversial
Slight narrowing of urethra distal to urethral sphincter on MCUG
May be soft narrowing on cystoscopy
Not thought to be obstructive